

ISOLATED TROCHLEAR NERVE PALSY ASSOCIATED WITH CAROTID–CAVERNOUS SINUS FISTULA

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SUMMARY

Cranial nerve ophthalmoplegia linked with a cavernous sinus lesion usually involves the third, fourth and sixth cranial nerve. Isolated fourth nerve palsy caused by carotid–cavernous sinus fistula (CCSF) is rare, and related case reports are sporadic in the literature. We report a 67-year-old woman with thunderclap-type headache and isolated right-side trochlear nerve palsy. The unique history and possible pathophysiologic mechanism are discussed. We propose that: (1) thunderclap-type headache could serve as a first symptom of CCSF; and (2) isolated trochlear nerve palsy within the cavernous sinus is not unusual. First-line clinicians should be alert and more aware of this entity. [International Journal of Gerontology 2009; 3(2): 129–132]

Key Words: carotid–cavernous sinus fistula, ophthalmoplegia, thunderclap headache, trochlear nerve palsy

Introduction

The manifestations of carotid–cavernous sinus fistula (CCSF) include ocular venous congestion, eyelid edema, chemosis, proptosis, pulsatile tinnitus, and various types of headache. Among these, the most noticeable symptoms are bruits and eye protrusion¹. Isolated trochlear nerve palsy within the cavernous sinus has been documented as the sole clinical finding. Because of the diversity of pathologic draining, isolated trochlear nerve palsy resulting from posterior-shunted CCSF is often misdiagnosed. Here, we present a female patient with right-side CCSF, whose first symptoms were thunderclap headache and right-side fourth cranial nerve paresis. The cranial imaging studies and characteristics of CCSF provided the diagnosis.

Case Report

A 67-year-old Taiwanese woman developed acute onset of vertical diplopia when looking to the left side. Her medical records showed that she had a history of hyperthyroidism and had an operation when she was a teenager; reasonable control and regular follow-ups were documented in her medical chart. One-and-a-half months before she was admitted into our ward, she started to suffer from headache. Initially, the headache was described as “thunderclap” in character and reached its peak within minutes. The pain was intermittent during the day and exacerbated at night. Each episode lasted between 2 and 3 hours. The patient reported aggravation when lifting heavy objects and during position changes. However, the headache did not worsen when coughing or sneezing. The headache ceased for 2 weeks. Unfortunately, the headache took another form, located at the right temporal area and radiating to the retro-orbital region. She went to a private clinic for help and over-the-counter painkillers were prescribed, but the pain was not alleviated. Blurred vision and tinnitus ensued later.



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Ophthalmic consultation diagnosed “double vision” instead of blurred vision by the cover–uncover test. Double vision worsened particularly when she turned her head to the left side. Other routine ocular tests were unremarkable.

Ear, nose and throat consultation showed that she had subjective pulsatile tinnitus in the right ear. The tinnitus was described as a “honing” sensation. Tympanography and auditory-evoked potentials were within normal limits.

She was thus referred to our department for evaluation. Clinical examination showed impaired interactive movement in the right eye. The Bielschowsky three-step test was compatible with right-side superior muscle weakness. The patient tilted her head to the left side in an attempt to compensate for double vision. There was no proptosis, eyelid swelling, conjunctival injection or audible bruits over the subclavian, neck, orbital or temporal areas. The remainder of the neurologic and physical examination appeared normal. Laboratory tests upon admission were insignificant.

A gadolinium-enhanced brain magnetic resonance imaging and angiography series elucidated a CCSF at the right cavernous sinus (Figures 1 and 2). Conventional angiography was arranged. It confirmed that the lesion

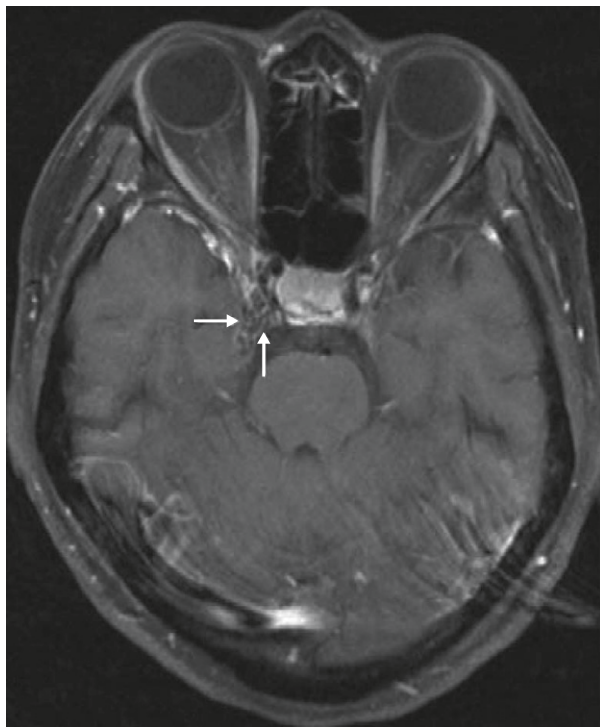


Figure 1. Contrast-enhanced T1-weighted image with fat suppression. Abnormal vessels around the right cavernous sinus (arrows).

was fed by meningeal branches of the internal carotid artery with flow fluxed posteriorly to the inferior petrosal sinus, then the internal jugular vein (Figure 3).

A neuroradiologist was consulted, and transarterial embolization (TAE) was suggested and performed (Figure 4). A Guglielmi detachable coil was then placed. Her diplopic symptoms greatly improved after TAE, but her pulsatile tinnitus remained. No other intervention was recommended. It was suggested that she had intermittent carotid compression. We followed-up using transcranial and extracranial sonography, and demonstrated no turbulent flow (Figure 5). She was symptom-free 8 months after embolization.

Discussion

CCSF is not an unusual neurologic disorder, but its clinical importance is usually overlooked². Clinical symptoms include ocular chemosis, tearing, proptosis, headache,

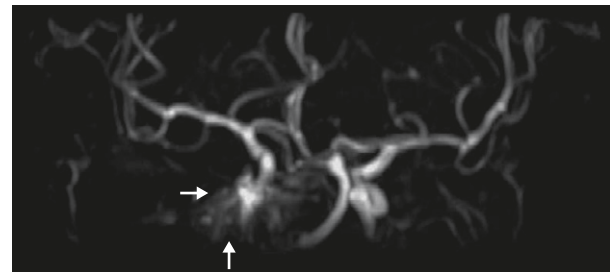


Figure 2. Intracranial magnetic resonance angiography. Abnormal vessels around the right cavernous sinus (arrows).

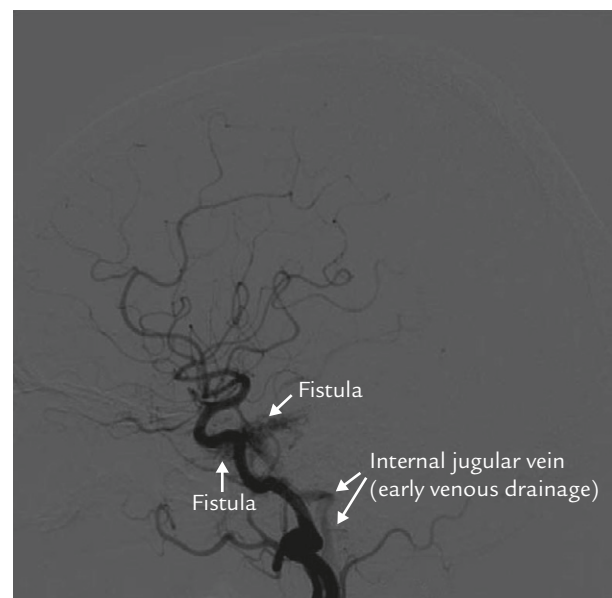


Figure 3. Pre-transarterial embolization state.

and tinnitus,³⁻⁷. Anatomically speaking, it could be subtyped into posterior and anterior drainage^{1,5,6,8}. Recognition of the anterior entity is not difficult, as its fistular flow is shunted into the superior ophthalmic vein. This often leads to orbital venous obstruction, and patients complain of “red eye”. Conversely, the posterior

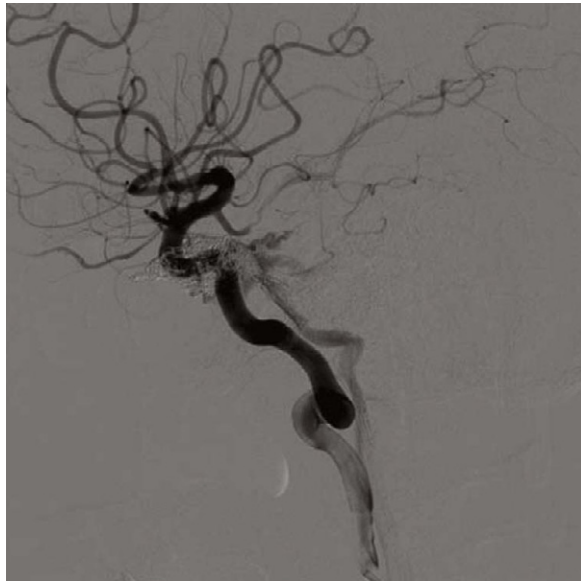


Figure 4. Post-transarterial embolization state. Coils placed in cavernous sinus via internal jugular vein.

CCSF usually lies in the posterior part of the cavernous sinus. Typical congested eye symptoms do not occur. Instead, the only clinical finding is often isolated/mixed ophthalmoplegia. Thus, compression/obstruction of the ophthalmic venous system is less prominent^{6,7,9}.

This patient presented to our department with simple diplopia and headache. The “thunderclap” headache, reaching its peak within minutes, had occurred before admission. On neurologic examination, the function of the right trochlear nerve was jeopardized. Infective factors were unlikely and initially ruled out in our patient on account of normal laboratory data and no infectious signs or symptoms. No traumatic history was taken. A vascular lesion or tumor was speculated. Thunderclap-type headache linked with obvious increased intracranial pressure can suggest subarachnoid hemorrhage, leading to the suspicion of an intracranial aneurysm compressing the nerve. Thus, an early imaging study is crucial. Localization is either in the supraorbital fissure or cavernous sinus lesion. A cavernous sinus lesion is more likely if there has been no accompanying oculomotor and/or abducens nerve palsy before imaging studies.

In our case, angiography proved to be a useful tool to diagnose a right-side CCSF which originated from the

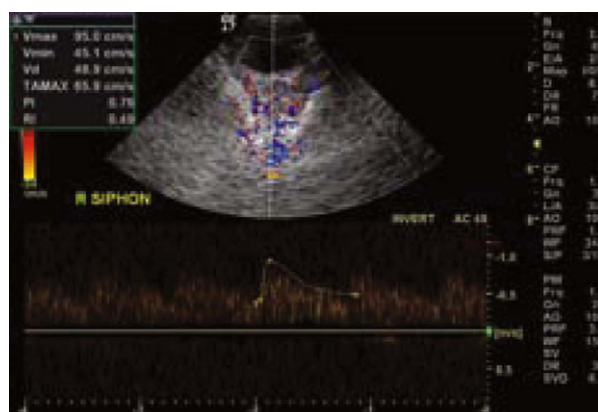
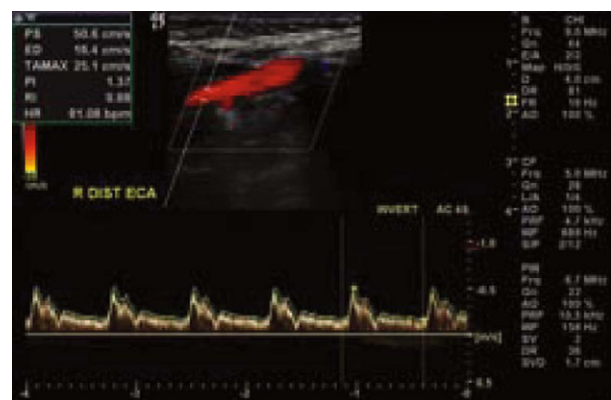
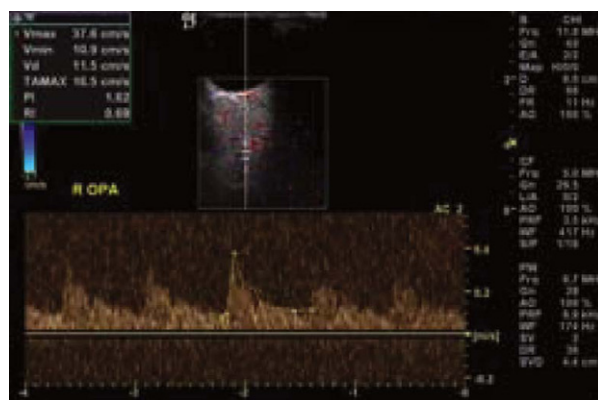


Figure 5. Post-transarterial embolization (TAE) follow-up. Right-side transorbital insonation (ophthalmic artery, siphon area) and extracranial artery showed a normal flow profile.

inferior petrosal sinus draining into the internal jugular vein, and not a subarachnoid hemorrhage derived from an aneurysm.

The literature suggests that the trochlear nerve enters the lateral dural wall of the cavernous sinus (i.e., between the oculomotor and the ophthalmic division of the trigeminal nerve) because of its close proximity to the tentorial incisura and its long course in the subarachnoid space. Any space occupying this region tends to have a bearing on neighboring structures^{8–10}.

Generally, in patients below 50 years of age, head trauma accounts for the majority of cases of trochlear nerve palsy. In contrast, vascular abnormalities are the leading cause in the elderly in regard to dysfunction of the fourth cranial nerve.

In the cavernous sinus, lesions that have been reported have varied from pituitary macroadenoma and meningioma¹¹ to herpes zoster⁴.

Carotid artery aneurysm compression⁹ is also an obvious contributing factor. Although extremely rare, a repeated form has been reported².

Isolated trochlear nerve palsy associated with CCSF is rare and is only reported sporadically in the literature^{1,3,8}. The nerve compromised within the cavernous sinus is typically accompanied by paresis of the oculomotor and/or abducens nerves⁶. The trochlear nerve is close to the oculomotor nerve in the cavernous sinus. Whether the mechanism, which affects the oculomotor nerve, can be applied to the trochlear nerve is still under investigation. Wu et al.¹² reviewed 33 patients with CCSF and found that the function of the oculomotor nerve was most vulnerable, followed by the abducens nerve, with the trochlear nerve being least affected.

The proposed mechanism of ocular nerve palsy caused by CCSF is different, and includes nerve compression by an expanding sinus¹, vascular steal, dural arterial steal, and venous congestion. Consensus is lacking, and the exact pathogenesis may be superimposed on individual anatomic variations. Therefore, the complexity of this condition is not homogeneous.

In conclusion, isolated trochlear paresis arising from a cavernous sinus lesion is not exceptional. A vigorous search for the cause, such as CCSF, is vital.

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